

Case 1

Demographics

Male
born 1974
185 cm
82 kg
BMI 23.96

SSc features

Diffuse SSc
Scl70+
ILD present
FVC 72% DLCO 53%
History Digital Ulcers
Upper GI involvement
Muscular involvement

Family History

Negative for CVD

Ongoing Treatment

MMF 1 g BID

Medical History

Chronic cystitis from 2019, Erectile dysfunction

Sildenafil 20 mg TID

CV Risk factors

None

Prednisone 5 mg QD

Esomeprazole 40 mg QD

Ranitidine 150 mg QD

Domperidone 10 mg BID

Symptoms

May 2018 presented with palpitations to A&E, NYHA II

Clinical Signs

Digital cyanosis

ECG: Atrial Flutter, 230 bpm, 1:1 conduction.

Holter ECG: 4118 SVEB, 1 SVT of 5 beats, 2420 VEB, 66 CPT, BIG 128, TRIG 63, 2 VTs (max 15 beats).

ECHO: normal morphology and diameters, global normo-kinesia, LVEF 52%, RA area 20 cm^2 , sPAP 32 mmHg, mild tricuspid regurgitation, normal pericardium.

LAB: CPK 110 U/L, ALT 33 U/L, TSH 8.57mU/L fT4 13.54 pmol/L, NTproBNP 318 pg/ml, TnI 62,4 ng/ml.

CMR: LVEF 49%, mild diffuse hypokinesia; RVEF 52%, normal kynesia; dilated RA 16cm^2 , mild-moderate TI, trivial pericardial effusion, no T2-STIR-BB alteration, T1-GRE-IR intra-myocardial LGE anterior-basal septum (mid-wall), infero-basal septum (non ischaemic pattern), diaphragmatic RV wall.

EMB: not done.

OTHER TESTS: none.

Therapy: Carotid sinus massage (not effective), Adenosine not effective, DC Shock effective → Carvedilole + Amiodarone.